Neuroectodermally Derived Neoplasm of Tonsil: Difficulty in Histomorphological Diagnosis

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Abstract Esthesioneuroblastomas are rare neuroectodermally derived neoplasms occurring in the upper nasal septum or cribriform plate. They have been rarely reported in other sites, when they are called ectopic esthesioneuroblastomas. Due to the scarcity of reports, publications and molecular/genetic studies of these tumours, their diagnosis is likely to be missed when they are ectopic. Similar to the case report in this journal by Zahedi et al., we had a case of neuroectodermally derived neoplasm of the tonsil, which was difficult to diagnose, due to the site of occurrence being unknown for these tumours.

Keywords Tonsil · Esthesioneuroblastoma · Paraganglioma · Neuroectodermal · Head and neck

Sir,

We read with enthusiasm the article “Ectopic Esthesioneuroblastoma of the Sphenoclivus: A Rare Entity”, by Zahedi et al. We agree that it is difficult to diagnose esthesioneuroblastoma in ectopic sites [1], especially when it is high grade.

We had a case of neuroectodermally derived neoplasm of the tonsil, where the list of differential diagnoses included esthesioneuroblastoma. But due to rarity of this tumour in ectopic sites and absence of molecular studies in these tumours, it was difficult to make a diagnosis.

Our patient was a 61 year old male, who presented with throat pain and swelling in the neck of 15 days duration. On examination, the patient had a mass in the left tonsillar fossa. A few bilateral cervical lymphnodes were found enlarged. CT scan of the neck showed an ill defined heterogeneously enhancing ulceroproliferative lesion in the left tonsillar fossa, measuring 19 × 19 × 10 mm. A few enlarged bilateral cervical lymph nodes were noted in the level III and level IV regions, with necrotic areas in the right level V region. A biopsy of the tonsillar mass was done, which with hematoxylin and eosin stains showed patternless sheets of polygonal pleomorphic cells with frequent mitoses (upto 19/10 high power fields) and necrosis. A differential diagnoses of poorly differentiated squamous cell carcinoma, malignant melanoma, neuroendocrine/neuroectodermally derived neoplasm and high grade lymphoma were considered, based on morphology. Immunohistochemistry that followed showed that synaptophysin (Fig. 1a) was strongly positive in the tumour cells. Chromogranin and NSE (Fig. 1b) were also positive. S100 highlighted a few positive cells focally, which were seen rimming tiny nests of neoplastic cells (Fig. 1c). Ki67 was positive in 51% of tumour cells. Cytokeratin (Fig. 1d), LCA, HMB45 and MelanA were negative. The differential diagnoses considered based on immunohistochemistry were malignant paraganglioma and esthesioneuroblastoma grade IV [2]. PET scan with 18F-FDG showed intensely hypermetabolic heterogeneously enhancing lesions in left tonsillar fossa and right level V cervical lymph node, both with an SUV max of 7.5. The patient’s blood pressure was 130/80 mm Hg, and plasma and urine metanephrines were negative. There was no family history of paragangliomas. Our patient was investigated for SDH mutation, which was absent.

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Fig. 1 Immunohistochemistry. Neoplastic cells stain positive with synaptophysin IHC ($\times 200$) (a) and NSE (Neuron specific enolase) ($\times 400$) (b). S100 stains sustentacular cells in the periphery of the

neoplastic cell nests ($\times 400$) (c). Cytokeratin is negative in neoplastic cells, and positive in the surface squamous epithelium ($\times 400$) (d)

Though ectopic esthesioneuroblastomas have been reported in the sphenoclivar region [1], pterygopalatine fossa [3], ethmoid sinus, nasopharynx, nasal lateral wall and floor [4], they have not been reported in the oropharynx or tonsils. Our case did not have any hypermetabolic focus in the upper nasal septum or cribriform plate on PET scan. Paraneoplastic syndromes such as inappropriate ADH secretion were not present [2].

As far as paragangliomas are concerned, there are no well-defined histopathologic features to distinguish benign from malignant paraganglioma, other than invasion and metastasis [5]. Our case, however, had many mitoses, 51% Ki-67 index and necrosis -features unusual for paragangliomas, including metastatic ones. The zellballen pattern was lost in some areas.

While paragangliomas are associated with mutations in succinate dehydrogenase subunits (SDH-B, -C, -D, SDH5), VHL, RET and NF1 [5], molecular/genetic studies in esthesioneuroblastomas have been few [2].

It is essential to differentiate between the two diagnoses since treatment and prognosis differ in these two

conditions. A diagnosis of malignant paraganglioma was concluded in our case because of the site of the tumour. The patient is being treated by the medical oncologist with chemotherapy.

Though, to the histomorphologist, differentiating between malignant paraganglioma and high grade esthesioneuroblastoma in the head and neck region would be a difficult task, the possibility of occurrence of either of them should be kept in mind. That will help in choosing the appropriate immunohistochemistry markers and molecular studies for correct diagnosis. Since the occurrence of these tumours in sites like tonsil is very rare, each and every case deserves to be highlighted; hence this case report. More reports and research regarding ectopic occurrences, genetic and molecular changes in esthesioneuroblastomas may reduce the under-reporting of this entity.

Compliance with ethical standards

Conflict of interest All the authors declare that he/she has no conflict of interest.

Ethical standards This article does not contain any studies with human participants or animals performed by the author.

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